CASE REPORT

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Imaging of an Intrahepatic Portosystemic Venous Shunt with an Associated Aneurysm: A Rare Entity

Divya Nijhawan, Kritesh Goel**, Utkarsh Garg**, Hardik Brahmbhatt***

Abstract

Background: Intrahepatic portal venous shunts are rare hepatic vascular communications between branches of portal veins and systemic veins. Early diagnosis is important because the condition can lead to hepatic encephalopathy and hypoglycaemia. Radiologists studying patients with liver disorders should be aware of this vascular anomaly and should also recognise that many occur in asymptomatic patients without liver disease and as such do not require treatment. In patients without any hepatic aetiology or a history of trauma, it is presumed to be spontaneous or congenital in origin.

Objective: The aim of our study is to describe the imaging findings in incidentally discovered asymptomatic intrahepatic portal venous shunts.

Key words: CT, intrahepatic portal venous shunts, liver, shunts, aneurysm.

Introduction

An intrahepatic portosystemic venous shunt is defined as communication between an intrahepatic portal vein and a systemic vein, including the hepatic and peri-hepatic veins, via an anomalous intrahepatic venous channel. Intrahepatic shunts between portal and systemic veins can be acquired (liver parenchymal diseases, post-traumatic, leaking of a portal vein aneurysm) or congenital[1]. Intrahepatic portosystemic shunts was first reported by Doehner *et al*[2] and later illustrated by Raskin *et al* in 1964[3]. The circumstances surrounding the discovery of an intrahepatic portal venous shunt in a patient without cirrhosis are variable. A few patients may present with hepatic encephalopathy due to high-output shunting. In this situation, hepatic dysfunction prompts an imaging examination and, thus, discovery of the shunt. In a series in Japan, of noncirrhotic patients with portal systemic encephalopathy, 36.2% of 47 patients presenting with encephalopathy had intrahepatic portosystemic shunts[4].

Intrahepatic portal venous shunts between a portal vein and a hepatic vein are much less common than those to perihepatic veins or the inferior *vena cava*. A 2003 report of angiographic findings suggests that only fifty patients have been reported in the English-language literature and that most of these cases (76%) were not associated with cirrhosis[5].

With the increased use of imaging studies, these lesions are likely to be increasingly encountered. We report the imaging features of incidentally discovered asymptomatic intrahepatic portal venous shunts in a 50-year-old female patient.

Case report

A 50-year-old female presented to the OPD with a short history of painful micturition with no relevant past history. On physical examination, vitals were normal with SpO_2 of 98% . Patient's blood pressure was 110/86 mmHg. Routine biochemical examination was unremarkable while urine culture showed many pus cells. Urea levels were raised (102 mg/dl). Serum ammonia levels were performed which were normal. There was no history of any kind of liver biopsy or any interventional procedure done.

Ultrasound of the abdomen showed normal size and echo-texture of the liver along with a large anechoic cystic lesion Color Doppler showed pulsatile flow within the veins adjacent to the right hepatic vein. The lesion showed venous flow in the color Doppler (Fig. 2). No e/o splenomegaly or ascites seen. Left kidney was echogenic and bulky.

Triple phase CT was performed. Patient received IV 150 ml of iodinated contrast material (iohexol; 300 mg I/ml). Contrast material was administered at a rate of 2 - 4 ml/s. Left kidney was found to be bulky and measured approx. 11.7 x 6.3 mm and showed reduced parenchymal enhancement (Fig. 1a). There was an incidental finding of focal collection of vessels in the anteroinferior segment of right lobe of liver (segment V) measuring approx. 38 x 34 mm showing enhancement in the venous phase with density equal to that of the blood vessels. The right hepatic

Junior Resident, Department of Radiodiagnosis, **Junior Resident, Department of Surgery, Maharishi Markandeshwar (Deemed to be) University, Ambala - 133 203, Haryana.

Corresponding Author: Dr Divya Nijhawan, Junior Resident, Department of Radiodiagnosis, Maharishi Markandeshwar (Deemed to be) University, Ambala - 133 203, Haryana. Phone: 8168558138, E-mail: dnijhawan5@gmail.com.

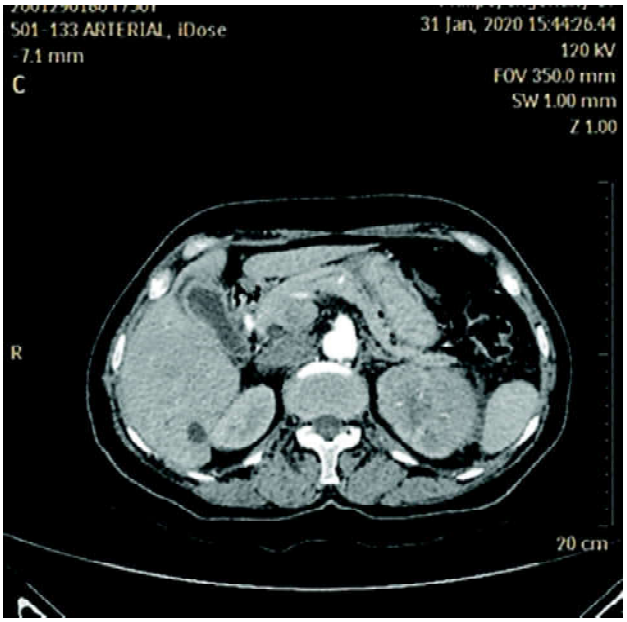

Fig. 1a: *Axial post-contrast CT (arterial phase) image shows a small hepatic cyst in segment VI of liver with a non enhancing area in segment V.*

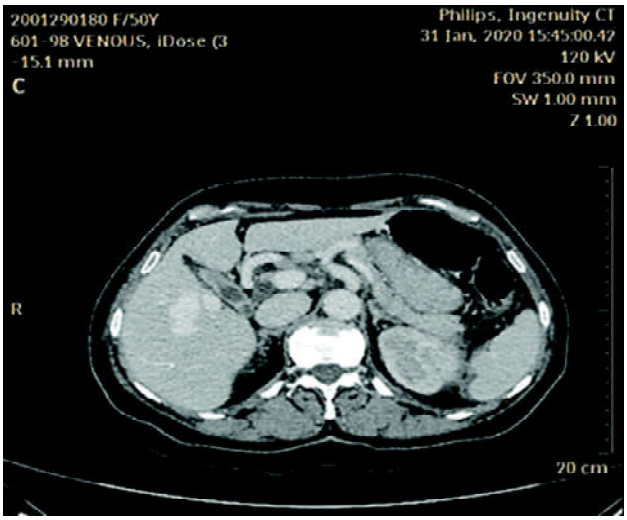

Fig. 1b: *Axial post-contrast CT (venous phase) demonstrating focal conglomerate vessels in segment V s/o intrahepatic portosystemic venous shunt between right portal vein and right hepatic vein.*

vein was dilated and measured approx 13 mm in calibre. Both anterior and posterior branches of the right portal vein were prominent with abnormal communication between the right portal vein branches and the right hepatic vein with resultant aneurysmal dilated vessels in segment V s/o porto-systemic venous shunt. Left branch of the portal vein was not dilated. No enhancement of the lesion was seen in the arterial phase. No other abnormal arteriovenous or venovenous shunt was seen. A small hepatic cyst measuring approx 11 mm was seen in segment VI. IHBR were not dilated. Final diagnosis of intrahepatic portosystemic venous

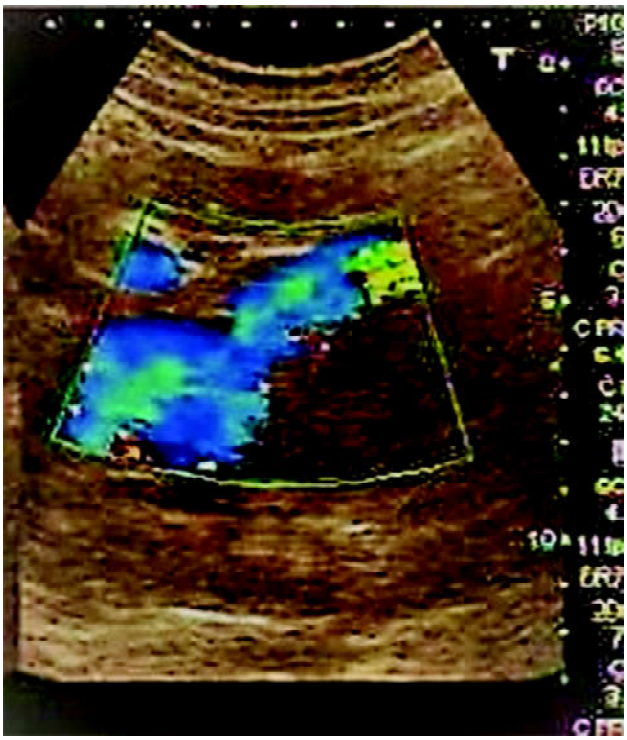

Fig. 2: *USG colour Doppler image showing venous flow in dilated cystic lesion adjacent to right hepatic vein showing venous flow on colour Doppler.*

shunt with aneurysmally dilated vessels (Type III Park's classification) was made.

Discussion

Portal-to-systemic venous communications are mostly seen on imaging studies in patients with portal hypertension from liver cirrhosis. These communications are largely extrahepatic and are commonly via the coronary vein, oesophageal varices, or retroperitoneal collaterals. Intrahepatic or transhepatic portosystemic communication refers to communication between the intrahepatic portal vein and a systemic vein[6]. They are much less frequent than extrahepatic shunts. Intrahepatic portal venous shunts were stratified by the system described by Park *et al*[7]. Type I is a single large tubular vessel of constant diameter that joins the right portal vein to the inferior *vena cava*; type II is a peripheral shunt in which solitary or numerous communications are found between peripheral branches of portal and hepatic veins in one hepatic segment; type III is an aneurysmal connection between the peripheral portal and hepatic veins; and type IV is multiple, diffuse communications between peripheral portal and hepatic veins in both lobes of the liver.

The origin of these shunts is the matter of differing views. When a 'portal vein-hepatic vein' communication is seen in

a patient without liver disease or a history of trauma, it is considered to be spontaneous or congenital in origin[8]. The basis of these shunts lies in the abnormality during the 4th week of intrauterine life in the development of vitelline veins and omphalomesentric system and the sinus venosus due to local absence of formation of sinusoids. Others speculate sudden rupture of a portal vein aneurysm into the hepatic vein is the cause[8]. During development, the right umbilical vein involutes and the left umbilical vein forms a direct communication with the ductus venosus (right hepatocardiac channel), bypassing the sinusoidal plexus of the liver[9]. Blood therefore flows from the placenta through the umbilical vein, ductus venosus, into the right hepatocardiac channel (later part of the inferior vena cava). After birth, the left umbilical vein forms the ligamentum teres and the sinus venosus forms the ligamentum venosum. Both the ligamentum teres and the ligamentum venosum are contiguous to the left hepatic lobe. Possibly these shunts represent persistent developmental communications[10].

Various embolising agents are used to occlude these shunts in symptomatic patients[5]. However in asymptomatic patients, no intervention is required as in our case[11].

Conclusion

The importance of this case was not only knowing the rarity of this pathology, but also highlighting the efficiency of imaging modalities like ultrasound and CT in detecting the condition. Cyst-like lesions in the liver should be evaluated with colour Doppler ultrasonography for flow characteristics and should be differentiated with triple phase CT.

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